37

Concerning Certain Non-traumatic Perforations of the Macula Lutea.

By G. E. De SCHWEINITZ, A.M., M.D.,

PHILADELPHIA, PA.

[*Reprinted from Transactions American Ophthalmological Society, 1904.*]

CONCERNING CERTAIN NON-TRAUMATIC PERFORATIONS OF THE MACULA LUTEA.

By G. E. de SCHWEINITZ, A.M., M.D.,

Philadelphia, Pa.

Traumatic perforation of the macula, or, to use the name which is employed in England, "hole in the macula," is a lesion to which in recent years a good deal of attention has been directed. Almost always it has followed the blow of some hard body, for example, a fragment of wood, a piece of stone, a bullet or a ball, which has struck an eye previously healthy. We are particularly indebted to Kuhnt,[1] Haab,[2] and M. F. Ogilvie[3] for studies of this affection, the last-named writer having analyzed fifteen cases and having reviewed the literature. A good many examinations indicate that the lesion under these circumstances is a fairly constant one, being confined to the central region, and characterized by a slightly punched out hole, either circular or oval in shape, with sharply marked edges and stippled bottom, crossed, it may be, with pigment lines. It is permanent, and may be noted years after the original blow has been struck. Sometimes the vision is surprisingly good, sometimes defective; sometimes a scotoma is demonstrable, sometimes it is not. According to Mr. Ogilvie, in a certain number of cases there is obvious detachment of the retina, while in others transparent retinal detachment is not demonstrable.

I give this brief description of this well-known affection to introduce the subject to which I would like to call attention, namely, that under certain circumstances we may find an exactly similar change in eyes that have not been subjected in any manner

[1] Zeitschr. f. Augenheilk., III, 1900, p. 105.
[2] Ibid, p. 113.
[3] Trans. Ophth. Soc. U. K., Vol. XX, 1900, p. 202.

18473-76

to traumatism. That such may be the case has not escaped the attention of that singularly acute clinical observer, Professor Haab. Describing perforations of the macula lutea after contusion of the eyeball, he remarks:[1] "It is probable that openings of this kind in the fovea may appear spontaneously in advanced age without any traumatism, probably owing to arteriosclerosis. In a case of this kind I saw such a perforation in both eyes in a woman sixty-four years of age, with marked arteriosclerosis and some albuminuria." Kuhnt, who first noted this type of macular change in 1891, but did not describe it in detail until 1900, records four examples of a peculiar change in the retina in the macula, for which he suggests the title *retinitis atrophicans sive rareficans centralis,* which corresponds to the description already given, namely, the presence of a red, exactly circular spot in the macula. Searching for an etiological factor, he found in one case trauma, in two cases nothing which could account for the condition, and nothing also in a fourth case, the patient having consulted him simply on account of reduced visual acuity. Two of his patients had reached such years (fifty-nine and sixty-one respectively) when the possibility of circulatory disturbances brought into existence by senile changes was suggestive. In this connection I wish to report the following case as illustrating the senile type of this affection:

Case I. *Macular Lesion exactly resembling the so-called Traumatic Hole in the Macula in a Patient the Subject of Chronic Heart Disease and Arterio-Sclerosis.*[2] — A woman, aged sixty-six, consulted Dr. Robert Saunders in April, 1895, in the hope of obtaining glasses for improving her defective vision, which in the right eye amounted to 6/12 and in the left eye about 6/60 eccentrically. The right eye, after the correction of a hyperopic astigmatism, regained normal vision, and was in itself without notable ophthalmoscopic changes. In the left eye Dr. Saunders discovered the appearances shown in the watercolor by Miss

[1] Atlas and Epitome of Ophthalmoscopy, Authorized Translation from the Third Revised German Edition, edited by G. E. de Schweinitz, A.M., M.D., Figure 47.

[2] This case has been briefly reported in the American Journal of Ophthalmology, Vol. XIII, 1896, p. 46.

Washington, which I exhibit, and brought the patient to me in consultation. The ophthalmoscopic conditions were as follows: The optic disc was circular and exhibited along its lower and outer border a broadened scleral ring, beyond which was a slight area of pigment disturbance. The papillo-macular bundle was a little paler than the rest of the surface of the disc. The blood-vessels were about normal in their distribution, but both systems somewhat smaller than is natural and the finer twigs markedly tortuous, as one is accustomed to see them in the subjects of arterio-sclerosis. Directly in the center of the macular region was an oval area of reddish color, its border being more sharply marked and of deeper tint than its center, in which were a few white dots. The halo which surrounded the macular region, that is to say, the macular reflex, was unusually distinct, and beyond it was a delicate fringe of faint pigment markings. The central lesion, or hole, appeared to be slightly deeper than the surrounding eyeground, the difference in refraction being half a diopter. Vision was unimproved by glasses. The peripheral visual field was normal, and there was a central scotoma five degrees on each side of the fixing point and three degrees above and below it. Physical examination revealed that the patient was the subject of chronic heart disease, with a well-marked aortic, obstructive murmur; for years she had been afflicted with rheumatism. I saw the patient only once, and it is my impression that Dr. Saunders told me that a subsequent examination of her urine proved this to be normal, although I have not made note of this in my case book. The superficial arterial system, radials, temporals, etc., gave the usual indications of arterio-sclerosis.

This form of macular affection is not to be confounded with localized changes in this region, as they occur in myopia (myopic central retino-choroiditis), or, for example, secondary to choroidal tears or hemorrhage as dark and light stipplings and spots, or as the result of nephritis, diabetes, anæmia, organic stomachic disease, etc., or finally, in ordinary senile macular disease in which there is a considerable area of yellowish-white spots interspersed with pigment dots and small round hemorrhages, or irregular

areas of erosions which may go on to atrophy of the elements and pigment heaping. It is probable that the lesion is one of the types of senile macular change to which Haab[1] made reference years before he especially investigated the traumatic perforations of the macula, and which he thought might account for the diagnosis of "Amblyopie ohne Befund" then often made in elderly patients, because the condition was not readily recognized except through a dilated pupil. His description is as follows: Usually smaller or larger, at times as large as the papilla, yellowish-red or whitish, or else darkly pigmented spots are evident which are confined solely to the territory of the macula, choroiditis elsewhere in the fundus not being present. I say the lesion which I here describe and picture, and which Kuhnt and Haab have also described, may belong to this class, but evidently it presents marked differences, already sufficiently detailed. It is, in short, a congener of the traumatic hole in the macula, and may therefore be called *non-traumatic perforation of the macula,* arising in the eyes of subjects of arterio-sclerosis.

Not only is this lesion liable to result from injury and to appear in the eyes of elderly subjects, but it may follow non-traumatic forms of irido-cyclitis. By way of illustration I cite the following case:

Case II. *Macular Lesion exactly resembling Traumatic Perforation of the Macula following Iritis.*[2] — A woman, aged fifty-five, in good general health except for occasional seizures of rheumatism, had a severe attack of influenza in January, 1895, followed two weeks later by severe bilateral iritis of the so-called serous type. The disease gradually yielded to treatment, and at the expiration of three months the visual acuity was in each eye 6/9. Pigment was present on the capsule of each lens, and there were floating vitreous opacities; otherwise no changes were noted. The patient was not again seen until nine months later, or just one year after the iritis had first appeared, when she returned with the statement that for three weeks the vision of the right

[1] Bericht der siebenter Ophthalmologisch Congress, Heidelberg, 1888, p. 429.

[2] This case has been partly deseribed in the Philadelphia Polyclinic, Vol. V, 1896, and in the Ophthalmic Record, November, 1898.



Macular Region of the Right Eye Containing Lesion Exactly Resembling Traumatic Perforation of the Macula Following Iritis.

eye had been noticeably failing, and that she was conscious of a scotoma in her visual field "looking like a plate with dark center and greenish edges." Examination yielded the following results: O. D. 4/60, O. S. 6/15. Exactly in the center of the right macular region was an oval, red-brown area, about one-third the size of the papilla, containing in its center two yellowish-white dots and a few fine white stipplings. This spot was surrounded by a greenish-white ring, somewhat raised, so that the red portion appeared as if at the bottom of a small pit, the sides of which were composed of the border just described. The macular reflex was unusually distinct. The accompanying watercolor by Miss Washington gives the appearance of this lesion. The optic disc, the retinal circulation and blood-vessels, and the general eyeground exhibited no abnormalities. The visual field was slightly contracted and contained a central scotoma. The opposite eye exhibited no similar conditions. Practically no favorable result was reached by treatment, although the various alteratives — mercury, iodide of potassium, arsenic, etc. — were tried in succession. A little more than a year after the appearance of this lesion, namely, on March 1, 1897, the spot was still distinctly visible and presented the characteristics already described, except that the greenish ring was no longer manifest, and the reddish part, which had occupied the center of the circle, was not quite so distinct.

The vision of the left eye remained as good as ever it had been after the original attack of iritis, namely, 6/12, until this date, when it began to fail, and became 6/45. There were some vitreous opacities, one large, semi-transparent one closely resembling a huge epithelial cell, but these opacities had been more or less constant ever since the original attack of iritis. On the 23d of the same month an area of macular degeneration exactly like that which occurred in the right eye began to be visible, and in a few days assumed the characteristics which belong to the one upon the opposite side, so that the illustration then made is an equally good picture of the present lesion. Vision, however, on this side never sank quite as low as it did on the other, and at the

last examination, about half a year ago, was 6/60, medication, as in the former instance, having had practically no effect upon the area of degeneration. The lesions were therefore exactly symmetrical.

When the patient was last seen, on October 24, 1901, or five and one-half years after the lesion appeared in the right eye and four and one-half years after it had developed in the left eye, with the exception of the absence of the greenish-white border and a somewhat greater expanse of the reddish areas, the conditions were unchanged. The accompanying watercolor, made from the left eye by Miss Washington, illustrates the state of affairs.

Evidently the lesions in this case exactly resemble the one previously recorded, and in turn are closely analogous to those which are caused by concussion injuries of the eye. The ophthalmoscopic picture is very like those depicted by Kuhnt and Haab, and somewhat like several of those given by Ogilvie, although in most of his the red color of the center of the spots is made darker than those which I present, being more nearly a blood color, and not the brick red which these watercolors indicate. Also in his cases there was more evidence of change, œdema, etc., of the immediately surrounding retina than in those at present under discussion.

In the absence of exact microscopic examination of such lesions, their pathogenesis is to some extent obscure. Naturally, hemorrhage in this region has been suggested; indeed, I adopted this explanation when I first described briefly my cases. But the entire behavior of these macular defects differs from that of those which are hemorrhagic in origin, chiefly in their unchanging character, lasting, as they do, without notable alteration for months and even years. Moreover, I had the opportunity of seeing one develop, beginning at first with great depreciation of vision, without distinct ophthalmoscopic change, to be followed by the gradual appearance of this oval or circular lesion. Circular macular hemorrhage, moreover, after iritis, in association with glaucoma and in arterio-sclerosis, presents totally different ophthalmoscopic appearances, as may be seen in the accompanying

watercolor, which, as time goes on, give rise to a spot of atrophy which includes all tissues to the sclera and is associated with surrounding disturbance of pigment lining.

Kuhnt, as may be inferred from the descriptive title which he suggests — *retinitis atrophicans sive rareficans centralis* — leans to the opinion that the condition represents a peculiar genuine retinitis of the posterior pole, which tends to central atrophy of its tissue. He points out that as the fovea is entirely devoid of capillaries and radial fibers, while the innermost portions of the macula possess them only in insignificant degree, the hypothesis is not far-fetched if one assumes that only those portions of the retina which are directly nourished by capillaries and bound together by Müller's fibers can withstand this inflammatory condition which tends to tissue disappearance, while the other portions are adapted to a melting away of their structure. Recently C. Harms[1] has described in great detail the microscopic lesions which he found in an eye of a seventy-seven-year-old man with senile macular changes — dark areas and light-colored spots. In general terms the lesions, consisting of atrophy and disappearance of the involved tissues, were confined to the macula and affected chiefly the neuro-epithelial layer. His measurements indicate that the lesion would have been clinically represented by a central scotoma about seven degrees in the horizontal and three degrees in the vertical meridian. He points out that such a condition does not correspond with those already found in central choroido-retinitis, but justifies Haab's assertion that there is a pure retinal senile affection.

In this conection the interesting observations of E. Fuchs[2] are important. He found in an eye blinded by irido-cyclitis following a blow by a piece of wood cavities in the outer reticular layer, partly filled with coagulated fluid, which communicated with similar cavities in the nuclear layers. Disturbances of the pigment epithelium and traces of previous hemorrhage were lacking; the choroid was practically normal; there was moderate round-celled infiltration of the iris and ciliary body. He regards the

[1] Klin. Monatsbl. f. Augenheilk, XLII, Bd. 1, 1904. p. 488.
[2] Zeitschr. f. Augenheilk., VI, 1901, p. 181.

condition as due to an inflammatory œdema, a transudation of serum from the retinal vessels, resulting from the low-grade inflammation of the ciliary body and iris. He has seen similar cavities in other injured eyes and thinks they account for the formation of "holes in the macula" after concussion injuries. Naturally the thought arises that similar cavities might explain the "hole in the macula" which I have described as a sequel of non-traumatic iritis.

Finally, it is to be remarked that degeneration of the ganglion cells, as it has been found by Ward Holden, Shumway, and others, in the macular changes in amaurotic family idiocy, has been suggested as an explanation of the affection I have been describing; indeed, I have myself advanced this hypothesis as a possible one. But the more one thinks of it the less suitable this theory becomes. The ophthalmoscopic signs are greatly different, not to mention the absence of any anatomical foundation for this idea in the examinations with the microscope which have thus far been made in analogous cases.

Evidently, then, the ophthalmoscopic appearances to which the term "hole in the macula" has been applied may arise from a concussion injury and then represent the usual type of traumatic perforation of the macula. Precisely similar lesions may follow an iritis or irido-cyclitis of non-traumatic origin and occur in the eyes of elderly persons, particularly if they are the subjects of arterio-sclerosis. Pathologically it would seem that the condition in so far as the last class is concerned depends upon a pure location of the disease in the macula, especially affecting the neuro-epithelial layer, and results in a form of atrophy or melting down of the tissues, the rest of the eyeground and the other coats of the eye being unaffected. Inasmuch as it has been demonstrated that traumatic irido-cyclitis and other injuries of the eye may be followed by a transudation of serum from the retinal vessels as the result of a low-grade inflammation of the anterior uveal tract and eventuate in cavities located particularly in the outer reticular and nuclear layers, a similar explanation is applicable to the "holes" which I have described as the result of non-traumatic iritis.

DISCUSSION.

Dr. C. J. Kipp. — I can add to the case of Dr. de Schweinitz one which differs somewhat from his — a young woman, who had never had iritis, nor cyclitis, nor any inflammatory disease. She had had for two years greatly impaired sight, and on examination the condition was found of which this is a representation. She had, beside this so-called hole in the macula, some pigment deposited in the disc itself; everything else was normal. She had a central scotoma, but the field was not contracted for white or colors. It shows that we can have the so-called holes in the macula without inflammatory conditions, or traumatism.

Dr. G. M. Gould. — I add an analogous case to the list, that of a patient whom I saw about six or eight years ago, a Japanese, who had central choroiditis with a pigmentary degenerative process arranged in concentric rings, looking as if made with India ink. There was this same punched-out perimetric hole in the macula. I could not determine that the man had ever had any specific disease, nor what was the cause of the condition. It puzzled me, and all I could do was through systemic treatment. I advised his going on a ranch out West. He could not, of course, read. In both eyes there was definite beginning formation of new maculas above. By putting on heavy prisms, bases down, I could get much better fusion with the apparently beginning new maculæ above. It seemed such an anomalous condition that I thought Dr. de Schweinitz might like to add it to his list.